CONGENITAL DEFECT IN THE ULNA.*

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THE condition of congenital absence of the ulna is one of the rarest anomalies to which the skeleton is liable. My patient also presented the extremely interesting feature of there being associated with it a true hyperplasia of bone in other portions of the body. A few of the numerous exostoses, from which she also suffered, are clearly shown in the accompanying X-ray photographs.

She first presented herself to me for treatment about two years ago, at the dispensary of the Howard Hospital, for what she believed was a dislocation of the right wrist, as the result of a traumatism and at that time I elicited the following history: M. D., aged 16 years, white, and by occupation an actress.

Family History.—Father living and well. Mother living but bed-ridden with "dropsy"; three brothers and three sisters living and well; four brothers and three sisters dead, two of diphtheria, one of meningitis and the others in infancy. Has no knowledge of any tuberculosis, malignant disease or syphilis in the family. No other member, immediate nor remote, of her family has suffered from any deformities.

Previous History.—Had diphtheria, scarlet fever, measles, chicken pox and typhoid fever two years ago, from all of which diseases she made a good recovery. Six months ago she noticed that exostoses were appearing, especially about the knees and ankles and the arms. Since that time they have steadily increased in size, and give constant pain which is increased by motion and pressure. She states that a "lump" appears whenever she receives a traumatism.

History of Present Condition.—Was playing with her sister who caught her by the right hand and gave it a violent pull,

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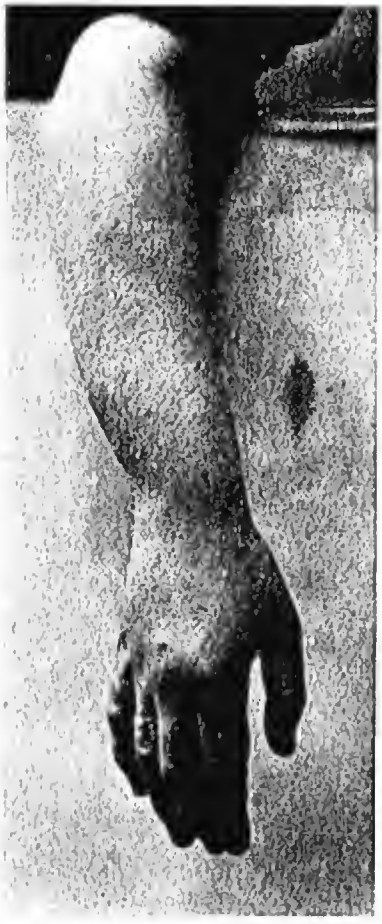
Fig. 1.



Absence of lower end of right ulna.

FIG. 2

Right forearm

Left forearm.

Congenital defect in ulna. Multiple exostoses.

which was at once followed by a marked deformity and complete loss of motion of the wrist joint. An attempt was made to make an examination without an anesthetic but the patient was very hysterical, so ether was given and the wrist joint was found not only freely movable in all directions, but the absence of the lower end of the ulna could be clearly felt. Further examination showed the presence of numerous exostoses. They are very marked on the upper end of the right humerus and on the left radius. The right wrist was dressed with a solution of lead water and alcohol and then put at rest upon a splint. X-ray photographs (see Fig. 1) were taken later that day and they show not only the absence of a portion of the ulna but also the exostoses on some of the other bones. Under rest the inflammation rapidly disappeared from the wrist joint and the patient soon had as good use of this arm as she had prior to the accident. I urged operation upon the exostoses, but the patient declined it, and I lost sight of her until she was admitted a year later to the Philadelphia Hospital, in the service of Dr. J. Chalmers Da Costa, and on June 13, 1906, his assistant Dr. Schwartz removed a number of the exostoses from the upper part of the tibia on both sides and from the external condyle of the femur on the right side. These were found to be very hard and attached firmly to the shaft of the bones. The patient made an uninterrupted recovery, and was eventually discharged from the hospital proper to the "Out Wards." She was readmitted to the surgical ward of the hospital in March, 1907, and on the 19th of that month she had the exostoses removed from the lower limbs. Two from the right knee, one from the outside and one from the inside. One from the outside of the left knee, the only one which was not firmly adherent to the femur. One also was removed from the lower end of the right fibula. The patient again made a good recovery and returned to the out wards. She has been there ever since except when admitted to the hospital, once for a gynecological operation and again for operation for appendicitis, both of which were successful.

Congenital absence of the ulna is a very rare condition; after a careful search of the literature I have been able to find but eight other reported cases. Förster, in his classical monograph, "Missbildungen d. Menschen," does not even mention

this anomaly, although he notes the frequent absence of the radius. The cases that have been previously reported are briefly the following:

1. GOLLER (quoted by A. Schnelle, "Inaugural Dissertation," Gottingen, 1875), in 1698, described a seven months' old foetus, in which both ulna with four fingers were absent, only the radii and both thumbs being present, and in the lower extremities only the tibia and great toes were present.

2. SENFTLEBEN (Virchow's Archiv., xlv, 1869, p. 303) notes the case of a recruit, aged 21 years, in whose left ulna there was a defect occupying the middle third of the bone. This defect measured two and one-half inches, and where the bone was absent a ligamentous band could be felt. The patient was in every other way normal.

3. ROBERTS (Trans. Path. Soc., Philadelphia, xiii, 1885, p. 4) reports the case of a man, 73 years old, whose right ulna was absent along with the third, fourth and fifth digits and their metacarpal bones. The pisiform, cuneiform and unciform bones were absent from the wrist. On the left side the ulna was present, but the third and fourth digits and the third metacarpal bone were wanting. The patient stated that his sister had one hand deformed like his and that she was the mother of a perfectly formed child. Another sister's child had a hand deformed like his right hand. The patient further stated that he had seven children, of whom three had malformations similar to his.

4. PRINGLE (Jour. of Anat. and Physiol., xxviii, 1893, p. 239) states that he had under observation, a man, aged 31 years, in whom both ulna were absent. There was no family history of deformities. The mother stated that the patient was born at full term, but during the early months of pregnancy she had received a severe fright. Right arm: The hand is provided with three fingers only, one well developed, which appears to be the middle finger, and two malformed ones, one to each side of the former. The wrist joint is freely movable, the trapezium is absent, but it is not possible to determine if any other carpal bones are wanting. There is no trace of the ulna. Left arm: The hand is narrower and less well shaped than the right; it also is provided with only three digits, the best developed of which seems to be the index. The wrist joint is freely movable, and the left trapezium is present. There is no trace of the ulna.

5. LANE (Trans. Clin. Soc., London, xxxii, 1898-9, p. 44) reports the case of a girl, 3 years of age, whose ulna consisted of two separate parts, whose pointed extremities slightly overlapped and whose axes varied considerably in direction. The lower end of the ulna was situated considerably above the level of the extremity of the radius. No evidence of any other deformity. It is interesting to note that this deformity was successfully corrected by the wiring in of the femur of a rabbit.

6. YUDT (Vrach. Gaz., S. Peterb., x, 1903, p. 342) notes the case of a woman, aged 62 years, who presented herself for treatment for a

fracture of the neck of the left humerus and in whom the right ulna was almost entirely absent. It consisted of a piece of bone, 7.5 c.m. long, extending downward from the elbow. The radius was arch-like in shape and somewhat thickened below where it articulated with the carpus; its upper extremity was dislocated forward and outward. The right hand was smaller than the left but all its bones were present. The only other deformity was an absence of the right pronator quadratus muscle. All the normal movements of the fore arm were well preserved except flexion of the elbow, which was much limited for purely mechanical reasons.

7. KACKKACHEFF (Russk. Gaz. S. Peterb., iii, 1904, p. 325) reports the case of a man, 24 years of age, in whom the left ulna was entirely absent. Pronation was well preserved. There was no other deformity.

8. AGAYEFF (Vrach. Gaz. S. Peterb., xii, 1905, p. 155) notes the case of a man, 40 years, who gave a history of polydaetylism in his mother, who had it in one of her feet, and in an uncle, on his father's side, who had it in both feet. The patient had had four children and all were perfectly formed. Right arm: The only deformity was polydaetylism of the little finger. Left arm: This arm was flexed at the elbow and in incomplete pronation. The hand had but three fingers, the thumb, index and middle. A rudiment of the little finger was also present. The humerus was normal and in the trochlear region was a round bony prominence, of the shape of the patella, movable in all directions and attached to the tendon of the triceps muscle. The ulna was completely absent, except for this rudiment. The little finger and the fourth and fifth metacarpal bones were absent, as was also the unciform and os magnum. The movements of this hand were somewhat limited, chiefly for mechanical reasons. No evidence of any other deformity.

The literature of multiple exostoses is indeed a voluminous one. I have gone through it with care and been able to find only one case where the hyperplasia of bone was associated with a congenital osseous defect. Battle (Trans. Clin. Soc., London, xxxix, 1905–6, p. 252) reports the case of a girl, 13 years of age, who had osteomata on nearly all the long bones, near their terminal epiphyses. The right ulna measured only six and one-half inches, while the left measured nine inches. The bone was well formed, only it was shorter than the radius, which had its shaft bowed outward and the head of the bone was displaced forward and outward.